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A CASE OF ICTERUS INFANTUM FROM CONGENITAL DEFICIENCY OF THE DUCTUS COMMUNIS CHOLEDOCHUS.[1]

BY

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THE following case, which I saw in consultation with Dr. Henry Moffat, of Yonkers, I am prompted to report because of the infrequency of occurrence of such cases and the very interesting character of the symptoms that were presented while the case was under observation.

For the data from which my notes have been compiled I am indebted to Dr. Moffat.

Mrs. P. gave birth, June 9th, 1887, to her second child, which was well formed, of average weight, and to all outward appearances physically sound.

Her labor was natural in every particular and of short duration. Nothing occurred to mar the mother's convalescence, and, so far as the infant was concerned, everything progressed satisfactorily up to the sixth day after its birth. The cord separated on the fifth day, leaving a clean and healthy surface. Up to the sixth day the child had nursed, its other functions were performed naturally, and nothing unusual was observed except an icteric tendency, increasing.

On the seventh day (June 16th), the infant refused the breast and became irritable. Soon it had a slight convulsion, which recurred at intervals of a few hours. On the morning of the eighth day (June 17th) Dr. Moffat was summoned and found the infant in a tetanic condition, its respiration had ceased, face was purple, eyes and mouth tightly closed, the upper and lower extremities were rigidly flexed, and the trunk and head were in a condition of opisthotonos. This state lasted for about twenty seconds when the natural condition of things gradually returned, with the respiration nearly normal and the pulse 120. The abdomen was tense, tumefied, and tympanitic. An enema was given which resulted in the expulsion of considerable flatus and

[1] Read before the Section on Practice of Medicine, New York Academy of Medicine, Nov. 15th, 1887.

apparently afforded the child some relief. In spite of appropriate medication, there was no improvement, but on the following day (June 18th) the child's condition seemed much worse; the temperature was 100.6, pulse 120–140, and the respirations accelerated and shallow. The jaundice was pronounced, and the convulsions recurred more frequently and were more severe in character. The abdomen was still tense, tympanitic, and tender. This condition of things continued until next day, June 19th (the fourth day of the attack and the tenth day of the child's life), when I saw the case with Dr. Moffat. I found the temperature 101°, pulse 130–140, jaundice very pronounced, abdomen expanded, tympanitic, and tender; a marked tumefied appearance about the region of the liver was observed, and on examination this organ was found greatly enlarged and sensitive to manipulation.

The nurse was directed to apply turpentine stupes, and eight grains of calomel were given. Nourishment was regularly administered by the mouth and rectum when necessary. Three free discharges from the bowels resulted in the night from the dose of calomel, and the next morning the condition of the patient appeared better. Toward evening, however, the temperature increased to 103°, pulse remained at 140, and the convulsions recurred frequently. A dose of one grain of acetanilide reduced the temperature to 100° in a short time, but the convulsions did not abate. An interesting circumstance was the total arrest of respiration after the convulsive attacks and the promptness with which the respiration was restored by artificial aid. A small amount of chloroform controlled the convulsions, but had no influence in lessening the frequency of the attacks. This condition continued without change until June 21st P.M. (the fifth day from the attack and the twelfth day of birth), when the child died of exhaustion. The character of the affection presented so many features of interest as well as obscurity that a necropsy was obtained. On opening the abdomen, the intestines were found coated with recent lymph and in some places were adherent. The liver was found greatly enlarged and engorged, and the gall-bladder was filled with blackish bile of a syrupy consistency. The hepatic ducts presented no abnormal appearances, but there was an evident constriction of the cystic duct and the common duct was impervious throughout its length. In the place of this duct there existed a fibrous cord-like band extending to the duodenum. When the chest was opened, the left lung was found completely collapsed and hugging close to the side of the spinal column in the posterior aspect of the thorax; the right lung was found in a natural state, fully expanded.

The case seemed so unique I determined to look up the literature of the subject and was rewarded by finding a very few cases reported presenting similar conditions. Eighteen or

twenty cases of various malformations about the gall-bladder or the several ducts were collected, but those reported by Campbell, Danforth, Glaister, Legg, Thommean, Murchison, Donop, and Maxwell Adams presented symptoms and post-mortem appearances very much like the case here reported.

As the following cases bear a relation to the subject, I will refer to some interesting portions of their respective authors' accounts of them, and believe that by so doing it will help to complete the record of all duct malformations of this class which have thus far been published.

J. Glaister[1] reported a case of icturus neonatorum in which a congenital stricture of the ductus communis choledochus was found on post-mortem examination. Jaundice appeared on the second day after birth, and on the third day the child developed "catches," spasm of the throat and contortions of the face. The respiration was characteristically Cheyne-Stokes, the pulse was slow and weak, the belly was tumescent, the skin dry, and the facial contortions increased in frequency and severity until death. Post-mortem investigation revealed a stricture of the common duct; the lungs were healthy in texture, the right was atelectatic, the left fully expanded.

Dr. Waring-Curran[2] reported a case of jaundice caused by recognized presence of a congenital lesion—spasmodic stricture—supposed to have resulted from an injury to the head, but did not cause death.

Dr. Lotze[3] recites the details of a case of congenital malformation of the liver ducts verified by necropsy.

A case of obstruction caused by "indurated cord-like plug of inspissated bile" was observed by Murchison,[4] who also alluded to a case reported by Lieutaud, where the obstruction proved to be a gall stone. In his lectures on liver diseases, page 375, a case of jaundice is spoken of, resulting in death, accompanied by epistaxis and ecchymoses. The lesion on necropsy was an obliteration of the common bile duct with perihepatitis.

Dr. Binz, of Bonn,[5] details two cases occurring in the same family, of children with obliteration of the ducts, but having

[1] London Lancet, 1879, vol. i., p. 293.
[2] Med. Press and Circular, Sept. 9th, 1868.
[3] Berliner klinische Wochenschrift, No. 30.
[4] Northern Jour. Medicine.
[5] Virchow's Archives, vol. xxxv., p. 360.

a gall bladder. In a third case, perihepatitis, with almost complete obliteration of the gall ducts, existed.

Dr. Binz mentioned two other cases [1] in which there was a rudimentary gall bladder and no trace of the biliary duct appeared.

In an old dissertation by Donop,[2] reference is made to a case in which the common duct was impervious throughout its length. Dr. J. N. Danforth [3] met with a case of entire absence of the ductus communis choledochus. The child had jaundice thirty hours after birth. Sixty hours after birth it began to nurse fitfully, would seize the breast eagerly and immediately forsake it with disgust. It was very irritable. The skin assumed a bronzed appearance in the place of a yellowish-brown color. Frequent attacks of vomiting occurred, which increased in severity until seventy-two hours after birth, when convulsions supervened and the child died in a state of profound coma. The liver was found enlarged and greatly congested. The gall bladder, occupying its normal position, was very much distended with bile of the consistency of syrup.

The cystic and hepatic ducts presented no unusual appearance, but were slightly enlarged. The common duct was distended and presented an abrupt termination without reaching the intestinal canal. His report was concluded with the remark that he had failed to find a similar case on record, though he had examined authorities within reach very carefully.

A. D. Campbell [4] gave an account of "two cases of icterus gravis infantum from deficiency of the cystic and hepatic ducts and one from plugging of the common duct." In the first case, the gall bladder was quite small and collapsed, and contained a little mucus which resembled gelatin in color and consistency. The bladder formed a closed sac having no outlet; the excretory ducts leading from the gall bladder and the liver were absent. The patient died without coma or convulsions. The second case, on post-mortem examination, revealed neither a gall bladder nor bile ducts, the liver was very much

[1] Virchow's collected works, p. 858. Clinical observations of Romberg and Hennock, p. 138.

[2] De Ictero speciatem neonatorum, 1828.

[3] Chicago Med. Jour., 1870, p. 110.

[4] North. Jour. Med., Edin., p. 237, vol. i.

enlarged, the venæ portæ, hepatic artery, and hepatic veins were all perfectly normal. After living six months, the child died of an attack similar to cholera, having been affected with incessant vomiting of a coffee-ground material up to the time of its death.

A male infant in the practice of Dr. Maxwell Adams died, on the eleventh day after its birth, of an obscure affection. On the third day the child manifested icterus mitis, but there was no apparent derangement of any function. On the seventh day, some oozing took place from the navel. The cord had separated on the sixth day, leaving a clean, healthy surface. On the eighth day a slight hemorrhage occurred which was controlled easily by ordinary means. Oozing of blood continued on the tenth day, but was quite inconsiderable, and no symptom of fever, spasm, or debility was present. On the eleventh day, Dr. Adams was summoned hurriedly and on his arrival found the child dead. No alteration had taken place in the child's symptoms until shortly before its death, when it refused the breast and sank, gradually, into a comatose state, expiring almost imperceptibly.

No trace of sloughing of any vessels that entered the liver from the funis was found on post-mortem examination. The iver was congested, but there was no trace of disease about the navel to account for the cause of the hemorrhage. The gall bladder contained a quantity of bile, which found no exit on account of an "indurated cord-like plug" of inspissated bile occupying the duct leading to the duodenum. The umbilical vein contained a clot about an inch in length.

J. W. Legg[1] reported an interesting case of a child who lived eleven weeks, and finally died of convulsions, from which it suffered for three days. Soon after birth it became jaundiced, and this condition progressively increased to the time of the child's death. On post-mortem investigation, the gall bladder was found shrunken, and contained only a small amount of yellowish fluid. The cystic duct opened without any winding into a cyst the size of a marble, situated to the right of the portal fissure, between the liver and the duodenum. The same cyst received the hepatic duct from the liver, and proved to be a blind sac with no passage into the duodenum.

[1] Trans. Path. Soc., London, 1876, xxvii., p. 178.

Numeley's[1] case of congenital obliteration of the hepatic ducts lived almost seven months. The child was admitted at the Children's Hospital suffering from jaundice, which had existed from its birth. A post-mortem revealed a small amount of yellowish serum in the abdominal cavity, but no peritoneal adhesions or recent lymph exudations were observed. The liver was firm, of a dark, blackish-green color. The orifices of the biliary ducts were not obvious when cut. The gall bladder was contracted, and contained only a little mucus.

D. Wilks[2] reported a case of obliteration of both cystic and hepatic ducts; the child lived six weeks.

Thommean[3] had a case of an infant that lived three months with an imperforate duct.

J. H. Morgan[4] reported a case of congenital malformation of the ductus communis choledochus.

Other malformations have been reported by Corrigan,[5] of a gall bladder communicating with the stomach, and cancerous tubercles about the pylorus. Freund,[6] "Ein Fall von congenitaler interstitieller Hepatitis mit Anomalie der Gallenausführungsgänge."

W. Gruber,[7] "Ein gabelförmig gespaltener Gallenblasenausgang. Ductus cysticus bifurcatus."

Herschl,[8] "Vollständiger Defect der Gallenwege beobachtet an einen 7-Monate alt verstorbenen weiblichen Kinde."

The above are all the cases of malformation of the liver ducts or gall bladder which I have been able to find recorded.

Whether or not the anomalies discussed are as rare as these few cases would naturally lead us to suppose, it is not possible to say. No doubt exists, I may say, that some cases are overlooked by the failure to procure autopsies of infants dying in a jaundiced condition soon after birth.

With regard to the case I have reported, the family history was good, and we have no theory to offer as to the cause of the phenomenon represented. I will add that the mother informed

[1] Trans. Path. Soc., London, 1872, xxiii., p. 152.
[2] Trans. Path. Soc., London, xiii., p. 119.
[3] Bull. Soc. Anat. de Paris, 1842, xvii., p. 52.
[4] Trans. Path. Soc., London, 1878, xxix., p. 137.
[5] Dublin Jour. Med., 1843.
[6] Jahrb. für Kinderh., Leipzig, 1875.
[7] Arch. für Path. Anat., Berlin, 1875.
[8] Wien. Med. Wochensch., 1865.

me that she was apprehensive during her puerperal period lest some harm should come to her or the child. The anxious state of her mind was occasioned by the loss of her first child, when a few months old, under very sad circumstances. Her mother also lost her two first-born in early infancy, one of them having died with symptoms of cholera infantum and jaundice.

These facts may be only coincidences in connection with the circumstances of the case herewith reported, but they will afford, nevertheless, a fruitful field for reflection with those who believe in the influence of external impressions on the production and development of fetal life.

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